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LETTER TO THE EDITOR

Contractile peripapillary staphyloma mimicking morning-glory disc anomaly

Dear Editor,

Although peripapillary staphyloma and morning glory disc anomaly are rarely encountered, accurate differential diagnosis based on disc appearance is essential because of their similar presentations [1]. However, few cases of contractile peripapillary staphyloma and contractile morning glory disc anomaly have been reported. This study describes a rare case of contractile peripapillary staphyloma that phasically mimicked a morning glory disc anomaly.

A 2-year-old Taiwanese boy presented with a crossed right eye since birth. His medical history was unremarkable. His ocular anterior segment was normal. The patient showed esotropia (ET) of 30Δ and a relative afferent pupillary defect in the right eye. Ocular motility was normal. Cycloplegic refraction was +0.75 + 0.75 × 30° in the right eye (RE) and +1.50 in the left eye (LE).

Ophthalmoscopic examination revealed a RE contractile peripapillary staphyloma (Fig. 1). Examination during contracted stage showed retinal vessels originating from the periphery of an excavated disc and coursing radially toward

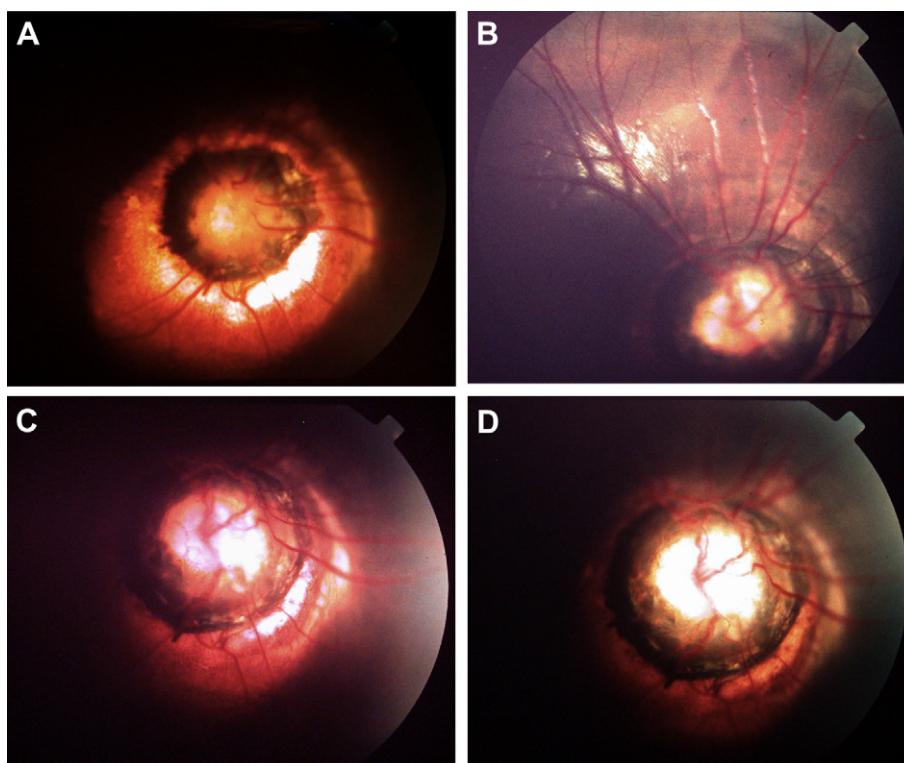


Figure 1. (A) Ophthalmoscopic images of contracted stage showing disc appearance resembling that of morning glory disc anomaly; (B, C, and D) image series of disc in expanded stage showing funnel-shaped peripapillary staphyloma.

the peripheral retina. A ring of hyperpigmentation surrounded the optic disc. A deep, funnel-shaped peripapillary staphyloma was noted in the expanded stage. The disc was at the bottom of the staphyloma, and the central trunk of the retinal vessels was visible. Brain magnetic resonance imaging (MRI) results were normal.

The patient was unavailable for examination for 5 years. At age 8 years, general examinations of the patient revealed normal development. Vision was 0.02 RE and 1.2 LE. During the 5-year interval, the patient had not consistently complied with the recommendation to occlude the left eye by wearing an eye patch. Cycloplegic refraction was $-4.25 + 2.50 \times 90^\circ$ RE and $+1.00$ LE. The ET was 20Δ , and no stereopsis was detected. Ophthalmoscopic examination revealed a mild dysplastic disc with posterior staphyloma but no pattern of contractility.

The embryogenesis of morning glory disc anomaly and peripapillary staphyloma is unknown. A case series study of patients with frontonasal dysplasia and basal encephalocele reported by Hodgkins et al revealed that all patients had either peripapillary staphyloma or morning glory optic disc abnormalities [2]. The authors suggested that these disc abnormalities have similar embryological origins. We speculate the morning glory disc anomaly observed in the current case resulted from a peripapillary staphyloma that did not adequately expand during embryogenesis.

Contractile cycles are reportedly related to respiration [3], light stimulation in the opposite eye [4], and transient visual loss [5]. Since morning glory disc anomaly is also associated with endocrine and central nervous system (CNS) anomalies, CNS imaging is essential [1]. The normal MRI results and normal development observed in the current patient suggested that the disease did not substantially affect other systems. Disc contractility also decreased as the patient grew older.

In congenital optic nerve diseases, visual acuity can range from 1.0 to complete lack of light perception. Although the current patient was treated with occlusion therapy in accordance with the literature, his final visual acuity was only 0.02. Together, the severely impaired vision, esotropia, and abnormal disc appearance suggested an underlying microscopic anomaly of retina/optic disc.

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Yu-Hung Lai

*Department of Ophthalmology,
Kaohsiung Medical University Hospital,
Kaohsiung Medical University, Kaohsiung, Taiwan*

*Department of Ophthalmology,
School of Medicine, College of Medicine,
Kaohsiung Medical University, Kaohsiung, Taiwan*

*Graduate Institute of Medicine,
College of Medicine, Kaohsiung Medical University,
Kaohsiung, Taiwan*

Gölge Acaroglu

*Department of Neuro-Ophthalmology and Orbital disease,
Ulucanlar Eye Hospital, Ankara, Turkey*

Hwei-Zu Wang

*Department of Ophthalmology,
School of Medicine, College of Medicine, Kaohsiung
Medical University, Kaohsiung, Taiwan*

Hsin-Tien Hsu*

*School of Nursing, College of Nursing,
Kaohsiung Medical University, Kaohsiung, Taiwan*

*Corresponding author.

100 Zih-You 1st Road, Kaohsiung 807, Taiwan
E-mail address: hthsu@kmu.edu.tw